**Case report**

Temporal arteritis in a young patient

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**ABSTRACT**

Temporal arteritis in the young is clinically and histologically different from classic giant cell arteritis of the elderly population. A male patient, aged 36 years, presented with headache and a nodule in his left temporal region. Histological examination of the nodule showed that the left temporal artery was encircled by a lymphoid tissue with prominent germinal centres. The arterial wall was infiltrated with mixed inflammatory cells, the internal elastic lamina was disrupted, and there was marked intimal hyperplasia. The patient was diagnosed with juvenile temporal arteritis. Because of persistent headache after surgical excision of the lesion, the patient was treated with prednisolone. Systemic vasculitides, classic giant cell arteritis, Kimura’s disease, and angiolymphoid hyperplasia with eosinophilia should be considered in the differential diagnosis of the disease.

**Introduction**

Vasculitis of the temporal arteries is rare in the young. To date, only about 40 cases under 50 year of age have been reported in the English literature. These patients can be divided into three groups (1); a) juvenile temporal arteritis (JTA), a localised eosinophilic vasculitis limited to temporal arteries, b) temporal arteritis secondary to systemic vasculitides such as polyarteritis nodosa (PAN) or Churg-Strauss syndrome, c) classic giant cell arteritis (GCA) occurring in the young. TA secondary to systemic vasculitides or classic GCA requires immunosuppressive treatment. However, JTA is a localised vasculitis, and surgical excision of the lesion is curative. Therefore, accurate diagnosis is important not only to avoid unnecessary medical treatment but also to reduce the economic burden of the disease (2). Herein, we describe a young male patient with TA and discuss differential diagnosis of the disease.

**Case report**

A 36-year-old Caucasian male presented with a 6-month history of bilateral temporal headache and a nodule in his left temporal region. The nodule was 2x1 cm in diameters, skin-coloured, and mildly tender on palpation (Fig. 1A). There was no history of trauma, major illness, or smoking. Headache was incompletely responsive to non-steroidal anti-inflammatory drugs. Physical examination was normal aside from the nodule in his left temporal region. Blood haemoglobin level and platelet count were within normal limits. White blood cell count (WBC) was 6,830 cells/μL, with mild eosinophilia (10.6% of total WBC count, normal up to 7%). Erythrocyte sedimentation rate (ESR) was 9 mm/h, and serum C-reactive protein (CRP) level was 3.34 mg/L (normal 0–6 mg/L). Blood chemistry panel and urine analysis were unremarkable. Hepatitis B surface antigen (HBsAg), anti-hepatitis C virus antibodies (anti-HCV), or anti-neutrophile cytoplasmic antibodies (ANCA) were negative. Serum immunoglobulin E (IgE) level could not be measured on admission. Colour Doppler ultrasonography of the nodule revealed that the superficial frontal branch of the left temporal artery was thickened and nodular in appearance. A hypo-echoic halo was observed around the arterial lumen. The arcus aorta and main branches were normal on magnetic resonance angiography. The involved segment of the temporal artery was excised. The artery was buried in a lymphoid tissue with prominent germinal centres (Fig. 1B). The arterial lumen was narrowed by markedly thickened intima (Fig. 1C). The adventitia, media, and intima were infiltrated with mixed inflam-
Differential diagnosis to be considered in the differential diagnosis of systemic inflammatory reactions, or connective tissue disease (i.e. fever, weight loss) are absent. ESR and serum CRP level are within normal limits. Eosinophilia may be present in some patients. The main pathological features include eosinophilic panarteritis with marked intimal hyperplasia and disrupted IEL. Granulomatous inflammation, fibrinoid necrosis, or giant cells are absent. Periarterial mononuclear cell infiltration with lymphoid follicles and germinal centres can be seen in some patients, and when accompanied by blood eosinophilia and elevated serum IgE level Kimura’s disease, which is a rare chronic inflammatory disease of unknown etiology that involves the subcutaneous tissue and lymph nodes, should be considered in the differential diagnosis (4). It presents as a soft tissue mass or lymphadenopathy in the head and neck region. Histological examination of the involved tissue shows eosinophil infiltration, lymphoid follicles with germinal centres, and proliferation of the small vessels. Whether JTA is a distinct disease or secondary to Kimura’s disease is controversial. In their first original report, Lie et al. stated that whether JTA represented a juvenile form of TA, a localised form of PAN, or Kimura’s disease was conjectural (3). Indeed, some authors have proposed that JTA is associated with Kimura’s disease (5, 6). The histological features in our patient were compatible with both JTA and Kimura’s disease; however, the clinical features were inconsistent with Kimura’s disease (i.e. soft tissue swelling or lymphadenopathy was lacking). Angiolymphoid hyperplasia with eosinophilia (ALHE) is another differential diagnosis to be considered (7). Small, superficial papules in the head and neck region, tissue eosinophilia, and lymphocyte infiltration and prominent vascular proliferation in the soft tissue are the main features of the disease (8). Periarterial lymphoid follicles are uncommon, and germinal centres are absent in ALHE. Ito et al. described a patient with JTA associated with hypereosinophilic syndrome (HES) (9). In our case, however, blood eosinophil count was <1,500/mm³, and none of the cutaneous findings of HES was present.

Discussion
Our patient had no systemic signs or symptoms suggestive of a systemic vasculitis. Moreover, histological examination of the biopsy material did not show findings consistent with GCA, such as giant cells or granulomatous inflammation in the arterial wall. A diagnosis of JTA was made with mixed cellular infiltration (including eosinophils) in the arterial wall, prominent intimal hyperplasia (IH), disrupted IEL, and absence of systemic involvement.

TA in the young population is different from classic GCA of the elderly in several aspects. First, TA in the young is extremely rare, with less than 40 reported cases to date. Second, it seems that these patients have a more favourable prognosis in terms of the ocular involvement when compared to classic elderly GCA patients. None of the reported patients had permanent blindness. Third, some patients (JTA subtype) do not need immunosuppressive treatment, and surgical excision of the lesion is seemingly curative in these patients.

JTA, first described by Lie et al. in 1975 (3), is the most frequent cause of TA in the young (less than 20 cases). Patients usually present with a painless or mildly tender swelling on the temple. The swelling can be unilateral or bilateral, and some patients report a history of head trauma. JTA is clinically and histologically different from both classic GCA and systemic vasculitides. Systemic involvement, systemic inflammatory reactions, or constitutional symptoms (i.e. fever, weight loss) are absent. ESR and serum CRP level are within normal limits. Eosinophilia may be present in some patients. The main pathological features include eosinophilic panarteritis with marked intimal hyperplasia and disrupted IEL. Granulomatous inflammation, fibrinoid necrosis, or giant cells are absent. Periarterial mononuclear cell infiltration with lymphoid follicles and germinal centres can be seen in some patients, and when accompanied by blood eosinophilia and elevated serum IgE level Kimura’s disease, which is a rare chronic inflammatory disease of unknown etiology that involves the subcutaneous tissue and lymph nodes, should be considered in the differential diagnosis (4). It presents as a soft tissue mass or lymphadenopathy in the head and neck region. Histological examination of the involved tissue shows eosinophil infiltration, lymphoid follicles with germinal centres, and proliferation of the small vessels. Whether JTA is a distinct disease or secondary to Kimura’s disease is controversial. In their first original report, Lie et al. stated that whether JTA represented a juvenile form of TA, a localised form of PAN, or Kimura’s disease was conjectural (3). Indeed, some authors have proposed that JTA is associated with Kimura’s disease (5, 6). The histological features in our patient were compatible with both JTA and Kimura’s disease; however, the clinical features were inconsistent with Kimura’s disease (i.e. soft tissue swelling or lymphadenopathy was lacking). Angiolymphoid hyperplasia with eosinophilia (ALHE) is another differential diagnosis to be considered (7). Small, superficial papules in the head and neck region, tissue eosinophilia, and lymphocyte infiltration and prominent vascular proliferation in the soft tissue are the main features of the disease (8). Periarterial lymphoid follicles are uncommon, and germinal centres are absent in ALHE. Ito et al. described a patient with JTA associated with hypereosinophilic syndrome (HES) (9). In our case, however, blood eosinophil count was <1,500/mm³, and none of the cutaneous findings of HES was present.

Fig. 1. (A) A localised swelling in the left temporal region, (B) structurally altered left temporal artery. It is buried in a lymphoid tissue with prominent germinal centres (H&E, x40), (C) arterial lumen is markedly narrowed by prominent intimal hyperplasia. There are mixed inflammatory cell infiltration and microvascular proliferation in the adventitia (H&E, x100), (D) disrupted internal elastic lamina (Verhoff’s stain, x100).
Treatment of JTA is surgical excision of the lesion, and recurrence has not been reported for up to 24 months of follow-up (3). However, it is clear that longer follow-up times are required to state that excision is definitely curative for the disease. Because our patient’s headache persisted after surgical treatment, he was started on prednisolone. He responded well and now asymptomatic and on low dose steroids.

The second form of the TA in the young is systemic vasculitides involving temporal arteries. Such involvement is extremely rare. PAN (10), Churg-Strauss syndrome (11), and thromboangiitis obliterans (12) are some systemic vasculitides involving temporal arteries in the young. The third, least common type is classic GCA. It is clinically and histologically similar to elderly GCA patients (13). However, blindness has not been reported in this group. Treatment of these patients should be tailored according to the underlying systemic vasculitides, for which novel therapeutic strategies and pathogenetic mechanisms have been proposed in recent years (14).

In conclusion, TA of the young is different from that of elderly patients. JTA subtype has the best prognosis, and surgical treatment seems to be curative. Short-term systemic steroid treatment could be considered in those patients who are unresponsive to surgical treatment. The other subtypes should be treated as systemic vasculitides.

References